Anterior Ischemic Optic Neuropathy in Eyes With Optic Disc Drusen

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Background: There have been anecdotal reports of anterior ischemic optic neuropathy (AION) occurring in eyes with optic disc drusen (ODD), but the clinical features of this condition have not been well characterized.

Objectives: To better describe the clinical features of AION associated with ODD and to compare the clinical features of this condition with those of "garden variety" nonarteritic AION.

Methods: We reviewed the medical records of 20 patients who experienced an episode of AION in an eye with ODD. In 4 patients, both eyes were affected; thus, 24 eyes were studied. The diagnosis of ODD was made by ophthalmoscopic identification, orbital ultrasonography, or computed tomographic scanning. We recorded age, sex, vascular risk factors, symptoms, visual acuity, visual fields, and results of the follow-up examination in all patients. These findings were compared with data from previously reported series of patients with nonarteritic AION.

Results: Our 20 patients included 14 men and 6 women (age range, 18-69 years; mean, 49.4 years). Vascular risk factors were identified in 10 patients (50%). Three patients reported episodes of transient visual loss before their fixed deficit. The visual acuity at the initial examination was 20/60 or better in 15 (62%) of the 24 eyes; 8 had a visual acuity of 20/20. The predominant pattern of visual field loss was an altitudinal or arcuate defect in 19 (79%) and a centrocecal scotoma in 5 (21%) of the 24 eyes. There was subjective worsening of vision before the initial neuro-ophthalmic examination in 11 eyes (46%) and objective documentation of progression in 7 eyes (29%). The final visual acuity was 20/40 or better in 13 (62%) of 21 eyes and 20/200 or worse in 3 (14%) of 21 eyes.

Conclusions: Our patients were strikingly similar to those with nonarteritic AION unassociated with drusen in regard to prevalence of vascular risk factors, pattern of visual field loss, and occurrence of a subsequent similar event in the fellow eye. In contrast, however, patients with ODD-AION were younger than those with nonarteritic AION, were more likely to report preceding episodes of transient visual obscuration, and enjoyed a more favorable visual outcome.


Optic Disc Drusen (ODD) are a relatively common congenital disc anomaly found in up to 2% of the population. They may appear as refractile bodies on the disc surface or may be buried and produce disc elevation. When buried, the main clinical significance of drusen is their resemblance to true papilledema. Impairment of visual acuity due to drusen is rare. In most cases, the visual deficit takes the form of slowly progressive field loss that may resemble glaucomatous visual loss and is often unnoticed by the patient. Occasionally, a more rapid decline of vision occurs, sometimes associated with decreased central vision. Some of these events are due to hemorrhage that may involve the disc, the peripapillary retina, or the vitreous. In other cases, visual loss is a consequence of vascular insufficiency affecting either the retina or the optic nerve.

Ischemia of the optic nerve in patients with ODD may or may not be associated with disc edema. In patients without acute disc changes, the presumed mechanism of visual loss is infarction of the retrolaminar optic nerve. Patients with acute disc edema are assumed to have anterior ischemic optic neuropathy (AION). The association of AION and ODD has been reported, but the clinical features of this condition are not well described.

METHODS

We reviewed the medical records of patients with ODD and acute visual loss examined in the neuro-ophthalmology clinics of the Mid-
RESULTS

Our 20 patients included 14 men and 6 women. Their ages ranged from 18 to 69 years (mean, 49.4 years). Of the 24 affected eyes, 13 were right and 11 were left. Vascular risk factors were identified in 10 patients (50%). Four patients had hypertension, 4 had diabetes mellitus, 1 had hypercholesterolemia, and 1 used tobacco. Other systemic conditions included melanoma, migraine, psoriasis, and pregnancy (1 patient with each condition). Other ocular disorders included chronic open-angle glaucoma in 2 patients, ocular hypertension in 1, previous iritis in 1, prior cataract extraction in 1, and congenital color blindness in 2. Visual loss was associated with mild eye pain in 5 eyes, described as an ache, pressure, sensation, or irritation. No patient had severe pain or pain with eye movement. In 4 eyes, visual loss was accompanied by a mild headache. Three patients reported transient obscurations of vision before onset of the fixed deficit. All patients were examined within 4 weeks of onset (mean, 8.5 days; median, 4.5 days).

Of 24 affected eyes, the visual acuity at the initial examination was 20/60 or better in 15 (62%); 8 had a visual acuity of 20/200. In 7 eyes, the visual acuity was worse than 20/200. Visual field loss was predominantly altitudinal or arcuate in 19 (79%) of the 24 eyes. Five eyes (21%) exhibited a centrocecal scotoma.

Patients described subjective worsening of vision before the initial examination in 11 eyes (46%) over 2 to 7 days. In 7 eyes (29%), serial examinations showed progression of visual loss within the first month following onset. A total of 14 eyes (58%) showed subjective or objective progression or both.

Follow-up information was available for 21 eyes, with an average interval between the initial and final examination of 2.9 years (range, 6 months to 16 years). Three eyes were not included in the assessment of visual outcome. In 2 eyes (1 patient), the patient was not available for reexamination. In 1 eye, optic neurotomy was performed and, thus, the outcome was not considered to represent the natural history of the disorder. At last follow-up, 13 (62%) of 21 eyes showed a visual acuity of 20/40 or better. Of the 21 eyes, 3 (14%) had a visual acuity of 20/200 or worse. Compared with the initial examination, 14 of 21 eyes showed no change or an improvement of 1 line or more. Seven eyes showed worsening by 1 or more lines. Of the 6 eyes that improved, the increase in Snellen acuity was 1 line in 1, 2 lines in 1, 3 lines in 1, 4 lines in 2, and 6 lines in 1. Of eyes that worsened, the change measured 1 line in 2, 2 lines in 3, 3 lines in 1, and 6 lines in 1. Of the 7 eyes that showed subsequent improvement of 1 to 7 lines.

The diagnosis of ODD was established by fundus examination in 7 eyes, by orbital ultrasonography in 15 eyes, and by computed tomography in 2 eyes. Ultrasonography and/or computed tomography were pursued in some patients because the disc appearance was suggestive of buried drusen (bumpy surface, scalloped margin, or deep peripapillary hemorrhage), in others because of the absence of vascular risk factors, in 2 because of unusually prolonged progression of visual loss, and in 1 because of recurrence of AION in a previously involved eye. Optic disc drusen were bilateral in 16 (80%) of the 20 patients. In 4 of these patients, an episode of AION was documented in each eye. In 2 patients, the interval between events was 3 months; and in 2 others, 6 months. In no patient were the 2 events simultaneous. In 2 other patients with bilateral ODD, the clinical findings were suggestive of a previous AION episode in the fellow eye. One patient experienced a second episode of AION in an involved eye.

PATIENT 1

A 44-year-old man experienced abrupt onset of spotty vision in the left eye, which was present on awakening. There was no associated head or eye pain, no systemic symptoms, and no history of focal neurologic deficit. His vision was subjectively worse the next day and then stable the day after. He was generally healthy and taking no medications.

A neuro-ophthalmic examination 3 days after onset of visual loss revealed a visual acuity of 20/20 OD and 20/300 OS. He identified all 15 pseudoisochromatic plates in each eye. There was a 1.8–log unit relative afferent papillary defect in the left eye. Goldmann perimetry in the right eye showed slight inferonasal constriction peripherally. In the left eye, there was a superior arcuate scotoma (Figure 1). A fundus examination showed marked elevation of the right optic disc without opacification of the nerve fiber layer and with no physiologic cup. The left disc was also quite elevated, had a bumpy surface, and showed,
in addition, capillary hyperemia and nerve fiber layer hemorrhages (Figure 2). A computed tomographic scan revealed calcification within the optic discs bilaterally, and was otherwise normal (Figure 3). The results of laboratory testing, including a complete blood count and the erythrocyte sedimentation rate, were normal.

On reexamination 3 weeks later, the patient reported no subjective change in vision but, in fact, visual acuity had improved to 20/25 OS, perhaps in part because the patient became more adept at extrafoveal fixation. He identified 10 of 17 pseudoisochromatic plates in the left eye, and the left relative afferent pupillary defect had decreased to 0.9 log units. Goldmann perimetry showed the same pattern of loss in the left eye, but with less central depression, including return of a small I-I-e isopter above fixation. The left disc was less edematous. All findings in the right eye were unchanged.

Two months later, the patient experienced sudden onset of painless visual loss in the right eye, which progressed during the first 48 hours and then stabilized. An ophthalmic examination 5 days after onset showed a visual acuity of 20/400 OD and 20/25 OS. Goldmann perimetry in the right eye showed a broad centrocecal scotoma (Figure 4). The right disc was swollen with peripapillary nerve fiber layer hemorrhages; and the left disc was pale, with drusen visible nasally (Figure 5). The results of additional testing for coagulopathy and systemic inflammatory disease were unrevealing.

PATIENT 2

This 59-year-old man experienced onset of painless visual loss in the left eye associated with mild photosensitivity. There was no history of headache or focal neurologic deficits. His ocular history was positive for bilateral ODD. His medical history included obesity and borderline hypercholesterolemia.

A neuro-ophthalmic examination 2 weeks after the onset of visual loss demonstrated a visual acuity of 20/20 OU, with normal color vision and a 0.9-log unit relative afferent pupillary defect in the left eye. Goldmann perimetry in the right eye showed enlargement of the physiologic blind spot and mild inferior depression. In the left eye, there was dense peripheral inferotemporal loss and a superior nasal scotoma (Figure 6). The fundus examination revealed marked bilateral ODD with peripapillary atrophy and anomalous branching of retinal arteries. In addition, in the left eye, there was mild thickening of the nerve fiber layer nasally with a few peripapillary hemorrhages (Figure 7). A fluorescein angiogram con-
firmed late staining of the left disc but not the right. The results of a complete blood cell count and the erythrocyte sedimentation rate were normal. Reexamination 2 months later showed mild worsening of the visual field in the left eye, with resolution of disc hyperemia and hemorrhages. Although there were no further episodes of acute visual changes, a reexamination 7 years later revealed further field loss nasally and superiorly in the left eye. 

Nonarteritic AION (NAION) is a common cause of acute visual loss. The mechanism in most patients is thought to involve a decrease in perfusion pressure within the optic nerve head that, in susceptible individuals, leads to infarction. This susceptibility may be related to abnormalities in the vasculature and the structure of the optic disc. The particular aspect of optic disc structure that has been implicated is a small cup-disc ratio, the so-called disc at risk. Optic disc drusen represent another form of disc anomaly in which the nerve head is crowded and cupless. Anecdotal reports of NAION occurring in eyes with ODD suggest an association between these 2 conditions as well. While a possible causative role has been proposed, other researchers have suggested that the coexistence of these 2 disorders is coincidental.

In this study, we analyzed the clinical features of 20 patients with drusen-associated AION (DAION). We hoped to provide a more complete description of this disorder and to clarify its relationship to the better-defined entity of “garden variety” NAION. Our 20 patients with DAION were a mean age of 49.4 years (range, 18-69 years). This contrasts with previously described populations of patients with NAION in whom the mean age was 63.4 years. Vascular risk factors were identified in 50% of our patients, compared with 60% of the NAION patients in the Ischemic Optic Neuropathy Decompression Trial. In addition to the primary symptom of acute monocular visual loss, 3 patients (12.5%) reported preceding transient obscurations of vision and 5 (21%) described mild ipsilateral eye pain. By comparison, 5.4% and 10% of patients with NAION reported these symptoms, respectively.

Four patients experienced bilateral sequential episodes of DAION, resulting in a total of 24 eyes studied. The visual acuity at the initial examination was 20/60 or better in 62% of the eyes (vs 31%-52% with this visual acuity in a combined NAION series). A visual acuity of 20/200 or worse was found in 29% (vs 35%-54% in the NAION series). The predominant pattern of visual field loss consisted of an altitudinal or arcuate defect in 79% and a centrocecal scotoma in 21%. In comparison, Repka et al found these patterns of visual field loss in 79% and 20%, respectively; Boghen and Glaser found altitudinal or arcuate defects in 71%, and Rizzo and Lessell reported a centrocecal scotoma in 26%.

Patients described subjective worsening of vision before the initial neuro-ophthalmic examination in 46% of episodes. In 29% of events, further visual loss was documented after the initial neuro-ophthalmic examination. These percentages are comparable to those reported in the Ischemic Optic Neuropathy Decompression Trial, in which 45% and 29% of eyes determined such subjec-
tive and objective progression, respectively. The visual acuity at the final examination was 20/40 or better in 62% of the eyes. This is in contrast to combined series of patients with NAION in whom this level of final visual acuity was found in 21% to 53%. A visual acuity worse than 20/200 was found in only 14% in our series (vs 31%-41% in the combined series).

Of 20 patients, 4 (20%) experienced bilateral sequential episodes of DAION. The incidence of bilateral NAION has varied widely in different series because of the nonhomogeneous patient population and differences in the duration of follow-up. The previously reported range of bilaterality is from 10.5% to 73%, with a recent estimate of 15% based on data from the Ischemic Optic Neuropathy Decompression Trial. In contrast, recurrent AION in a previously affected eye is extremely uncommon. Only 1 of 21 eyes with greater than a 6-month follow-up experienced a second episode of AION, occurring 10 years after the first event. Recurrent NAION has been reported in fewer than 5% of patients.

Based on these observations, many of the features of AION associated with ODD are strikingly similar to those of standard NAION. Specifically, the prevalence of vascular risk factors, the patterns of visual field loss, and the frequency of progressive visual loss are all comparable in the 2 groups. This suggests that the pathogenesis of visual loss in DAION may be similar to that of NAION. Is there a common denominator in these 2 conditions that might explain such a similarity? Both conditions have been associated with a small scleral canal. This anatomical configuration has been accepted as a factor in the pathogenesis of NAION because of axonal crowding with secondary vascular compromise. Such crowding may similarly predispose to AION in eyes with drusen.

In contrast to the above similarities, we found some differences in our patient group compared with previously reported series of patients with NAION. While the age range of our patients was broad, the mean of 49.4 years was considerably lower than the 63.4 years reported for patients with NAION. One possible explanation for this difference is that the drusen bodies themselves act directly on healthy optic disc vessels to cause infarction. Another possible explanation for this age difference might be related to the progressive thinning of the nerve fiber layer that occurs in eyes with ODD. Over time, the optic disc becomes less crowded and, thus, may no longer be susceptible to disc infarction. In other words, if a patient with ODD reaches the age of 60 years without experiencing a disc infarction, the likelihood of doing so thereafter may be small. In support of this explanation, we found that only 3 of our 20 patients with ODD experienced an episode of AION after the age of 60 years. Finally, the age difference could reflect a sampling bias because of more frequent imaging studies in young patients with AION.

Another contrast between our patient group and patients with NAION without drusen is the difference in overall visual outcome. Almost two thirds of involved eyes in our series had a final visual acuity of 20/40 or better, and only 3 were left with a visual acuity worse than 20/200. This more favorable outcome may reflect the younger age of our patient group. The optic disc in a younger individual may have more capacity for autoregulation and collateral circulation than that of an older patient. In support of this concept, we found that our 3 youngest patients, aged 18, 26, and 38 years, had final visual acuities of 20/30, 20/30, and 20/400 in the involved eye, respectively, compared with our 3 oldest patients, aged 62, 65, and 69 years, who had final visual acuities of 20/60, 20/400, and 20/50 in their involved eye, respectively. The visual acuities at onset were also better in our series compared with those of NAION patients without drusen, but such differences among studies are difficult to interpret because of variability in the timing of the initial examination.

We also found a greater frequency of transient obscurations of vision in our patients compared with those with standard NAION. While transient visual obscurations are well described in patients with ODD, they are quite uncommon in patients with NAION. A history of transient visual obscurations in a patient with NAION should, thus, raise the possibility of underlying drusen. Although we noted a higher frequency of pain in our group, the discomfort in all patients was mild, de-

Figure 7. Patient 2. Fundus photographs. A, The right disc shows extensive drusen with peripapillary atrophy and anomalous branching of retinal vessels. B, The left disc shows similar findings but with acquired disc edema nasally and a deep retinal hemorrhage at the inferonasal disc margin.
scribed as a pressure or an ache rather than actual pain, and was self-limited in all. The significance of this observation is unclear. We considered the possible alternative diagnosis of optic neuritis in these individuals, but this seemed unlikely on clinical grounds.

In summary, our study supports a real (nonincidental) association between ODD and AION. Patients with DAION are generally younger than 50 years, younger on average than patients with standard NAION. In most other respects, their clinical features are quite similar to those of patients with standard NAION, including the prevalence of vascular risk factors, the pattern of visual field loss, and the occurrence of a similar event in the fellow eye. In most patients, the visual outcome is more favorable than in patients with NAION without drusen, and the likelihood of a recurrent event in an affected eye, as in patients with standard NAION, is low. Patients can be counseled accordingly.

This condition may be more common than previously appreciated. The optic disc in the fellow eye of a patient with NAION typically appears crowded and full. In some individuals, this appearance may reflect underlying (buried) disc drusen rather than simply the typical NAION disc at risk. Drusen that are located more posteriorly (near the lamina cribrosa) may be more likely to cause visual loss than those on the disc surface. It is these deep drusen that are more difficult to detect on an ophthalmoscopic examination. Moreover, the disc elevation characteristic of buried drusen in young individuals is less prominent in older patients in whom progressive axonal dropout has occurred, thus making the diagnosis of drusen more challenging. A prospective study using ultrasonography in patients with NAION would furnish a better assessment of the actual frequency of drusen in patients with this disorder.

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